Large Organized Intrapericardial Hematoma Presenting as a Case of Constrictive Pericarditis and Superior Vena Cava Syndrome

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Abstract:
Superior vena cava syndrome and constrictive pericarditis are rare complications of intrapericardial hematoma. We herein report a young patient presenting with a mass attached to the heart causing pressure on the right atrium and superior vena cava, and leading to superior vena cava syndrome and constrictive pericarditis. The exact pathogenesis was not determined but possibly was related to pericardiocentesis 29 years prior to presentation. Diagnosis was established at surgery and confirmed by histopathology. Resolution was partial because complete resection was not possible due to adherence of the mass to the right ventricular wall.

Keywords: Superior vena cava syndrome, constrictive pericarditis, pericardial hematoma

Introduction:
Superior vena cava syndrome (SVCS) results from interruption of the venous return of blood from the head, upper extremities and thorax to the right atrium. Today in modern era, most cases of SVCS are caused by malignant tumors of the mediastinum, mostly lung cancer and lymphoma. Less common causes include fibrosing mediastinitis, thrombosis of indwelling central venous devices and/or pacemaker leads and intrapericardial hematoma (either traumatic, complications from dissecting aortic aneurysm or following heart surgery) compressing the right atrium. Coagulated blood within the pericardial space has been reported to cause a condition mimicking constrictive pericarditis. The manifestations of SVCS and constrictive pericarditis due to intrapericardial hematoma may appear after weeks or months and in certain instances, can be delayed by several years. We herein report a case of large organized intrapericardial hematoma presenting as superior vena cava syndrome and constrictive pericarditis, and review pertinent literature.

Case history:
A 37-year-old male, Indian patient was admitted to Hamad General Hospital with a one month history of progressively worsening exertional dyspnea associated with palpitation and fever mostly at night. He denied any complaint of cough, chest pain, or weight loss. His past history was unremarkable except for pericardiocentesis at the age of eight for an unknown cause. He denied smoking, alcohol consumption or drug abuse and had no history of trauma or accidents. Physical examination revealed a thin patient with a temperature of 36.5°C, blood pressure of 110/70 mm Hg and pulse 70/minute. Neck examination revealed markedly engorged neck veins and raised JVP (Figure 1). Other physical findings were right basal crepitations on chest auscultation, a visible apex beat in the 5th intercostal space with loud first heart sound, visible abdominal veins, distended abdomen with ascites and hepatomegaly. Investigations revealed a white blood cell count of 6200/mm³, hemoglobin at 11.3 gm/dL, platelets at 124,000/mm³ and normal liver and kidney function tests. Chest radiograph revealed globular enlarged cardiac shadow, right midzonal infiltrate and pulmonary congestion with pericardial calcification.

Figure 1: Photograph of the neck of the patient showing markedly distended non-pulsatile neck veins.
Electrocardiogram showed atrial fibrillation. Transthoracic echocardiogram revealed global hypokinesia with an ejection fraction of 45% and an extracardiac echogenic shadow pushing the left ventricular wall from the posterolateral side. A computed tomographic scan of the chest showed a calcified tubular mass extending from the right upper part of the pericardium, curving anteriorly, and reaching the left side causing pressure on the left ventricle and early reflux of contrast to the inferior vena cava. Also seen was a scar in the apex of the right lung with calcification in the left lower pleura, spleen and a small calcified mediastinal lymph node (Figures 2A and 2B). Ultrasound of the abdomen showed an enlarged congested liver with a span of 16 cm, prominent hepatic veins and superior vena cava with splenomegaly and ascites. Purified protein derivative (PPD) was positive. Sputum for acid-fast bacilli smear and culture was negative on three occasions. The patient was treated initially with intravenous furisemide and digoxin to control atrial fibrillation and pulmonary congestion. He then underwent thoracotomy and cardiopulmonary bypass. Partial pericardiectomy was performed to remove thick, fibrous pericardium encasing the superior vena cava, right atrium and inferior vena cava. A mass with a heavily calcified wall was found around the right atrium, right ventricle and left ventricle. Partial excision of the mass was made because it was attached to the right ventricle wall. The wall of the right ventricle was very thin and had to be repaired. Histopathology of the excised mass showed extensive fibrosis, calcification organizing hemorrhage, cholesterol clefts and minimal chronic inflammation. No evidence of granuloma or malignancy. The findings were consistent with organizing hematoma. A post-operative course was uneventful however he continued to have residual distension of neck veins. After discharge from hospital he was seen in the outpatient clinic on a few occasions but follow-up visits failed as he left Qatar for his native country.

Discussion:

The superior vena cava is formed by the union of the right and left brachiocephalic veins and empties into the superior-posterior right atrium. It is the major draining system for blood returning from the upper extremities, head and neck, and upper torso. The superior vena cava is relatively thin-walled and lies within undistensible space in the mediastinum, making it susceptible to external compression. In a recent review by Rice TW et al (2), of the 78 cases of SVCS seen over a five year period, tumors accounted for 60% of cases, indwelling intravascular device for 28%, fibrosing mediastinitis for 8%, hematoma due to aortic dissection for 1% and all others cases for 3%. Intrapericardial hematoma compressing the right side of the heart, especially the right atrium, may present with features of superior vena cava syndrome and pericardial constriction. Most reported cases of intrapericardial hematoma presenting as SVCS and constrictive pericarditis were reported in association with aortic dissection, (6) cardiac surgery (3, 4) and following chest trauma. (5)

Intrapericardial bleeding may present acutely causing cardiac tamponade. However, if pericardial hematoma develops and persists, it will induce intrapericardial inflammation, granulation tissue formation, adhesions and calcification leading to constriction. (7) In the chronic form, the patient may present after a latent period with breathlessness, chest tightness, ankle swelling, cyanosis, facial and neck swelling with elevated jugular venous pressure and lower limb edema. (7)

In the present case, the diagnosis of intrapericardial hematoma as a cause of the patient's symptoms was not seriously considered pre-operatively. A mass of malignant or inflammatory origin attached to the heart was suspected, however diagnosis was established only post-operatively. The presence of extensive fibrosis, adhesions...
and calcification upon histological examination indicated chronicity. Our patient was young with no recent history of dissection, myocardial infarction or chest trauma to explain the occurrence of the intrapericardial hematoma. However, he gave a vague history of pericardiocentesis at the age of eight for an unknown cause. Pericardial constriction has been reported to occur up to 20 years after chest trauma\(^5,8\) but the history of pericardiocentesis in our patient was 29 years before presentation. Whether this incident was related to his current problem is not known. Blood leak into the pericardial cavity through the very thin walled right atrium could be another explanation however, it is important to emphasize that in two-thirds of pericardial hematomas, no identifiable cause is found and therefore they are labeled as idiopathic.\(^8,9\) Chest radiographs, CT scans, MRIs and echocardiograms help in making diagnoses. A chest radiograph may show pericardial calcification while a CT scan and MRI more precisely identify the hematoma and the presence of calcification.\(^7-10\) In our patient, the CT scan and echocardiogram demonstrated the presence of the pericardial mass but it was not until a thoracotomy was completed, and the biopsy result of the excised mass was available, that the diagnosis of intrapericardial hematoma was established. Cardiac catheterization helps in making the diagnosis of cardiac constriction by demonstrating equalization of diastolic pressures in all chambers, with a typical dip and plateau pattern of right and left ventricular diastolic pressures. Unfortunately, catheterization was not performed in our patient. Surgery is the definitive treatment\(^7\) and it may take the form of simple evacuation or stripping of the epicardium.

Although the outcome in most reported cases of SVCS and pericardial constriction due to intrapericardial hematoma was frequently fatal,\(^6,7\) the outcome in our patient was favorable with resolution of the symptoms of shortness of breath, cough and chest pain, and improvement in the ejection fraction on the echocardiogram. He continued to have residual signs of superior vena cava obstruction in the form of distended neck veins and echocardiographic features of constriction. Their persistence is most probably related to the partial resection of the mass because it was very adherent to the heart and superior vena cava. The explanation of the poor outcome in the reported cases reflects that most of these cases were seen in association with aortic dissection which usually carries a worse prognosis.

**Conclusion:**

Superior vena cava syndrome and pericardial constriction due to intrapericardial hematoma is rare, with most cases occurring in association with aortic dissection and occasionally following trauma. Keeping a high index of suspicion and the use of available diagnostic modalities of echocardiography, CT scan, MRI and cardiac catheterization helps in early diagnosis. Treatment is surgical either by evacuation or pericardial stripping.

**References:**